

Pericallosal Lipoma and Agenesis of Corpus Callosum with Associated Lipoma of Choroid Plexus: A Rare Case Report

Qays A. Hassan

Qays A. Hassan, Radiologist specialist, Section of Radiology, Department of Surgery, Al-Kindy College of Medicine, University of Baghdad, Baghdad, Iraq

Correspondence to: Qays A. Hassan, Radiologist specialist, Section of Radiology, Department of Surgery, Al-Kindy College of Medicine, University of Baghdad, Baghdad, Iraq

Email: qtimeme@yahoo.com

Telephone: +9647722604163

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ABSTRACT

Pericallosal lipomas are rare congenital malformative lesions occurring in the interhemispheric fissure closely related to the corpus callosum which is usually abnormal. Pericallosal lipomas may extend into either or both lateral ventricles and appearing as lipomas of choroid plexus in the absence of corpus callosum. I observed this in an interesting case, and, as this is uncommon, I reporting it in order to increase awareness about this unusual presentation and to review briefly criteria of its pathogenesis & diagnosis.

Key words: Intra-cranial lipomas; Lipoma of choroid plexus; Corpus callosum; MRI

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INTRODUCTION

Pericallosal lipomas constitute almost half of all intracranial lipomas and are commonly associated with corpus callosal anomalies^[1].

Pericallosal lipomas may continue into the lateral ventricles, which is a very rare presentation^[2,3]. Although pericallosal lipoma is often described as corpus callosal lipoma, it is most often pericallosal in location. The pathogenesis of a pericallosal lipoma is considered to be the result of an anomalous persistence and differentiation of the mesenchymal tissue of meninx primitiva into mature lipomatous tissue during the development of the subarachnoid cisterns in the first trimester of pregnancy^[2].

Pericallosal lipoma can be clearly classified into two distinct types having different imaging morphologies and associated brain anomalies. One type consists of anteriorly situated round or cylinder-shaped lipomas, termed tubulonodular. These lipomas are generally measuring greater than 2 cm in diameter, have a high incidence of corpus callosum dysgenesis and fronto-facial anomalies and can extend into the choroid plexus/lateral ventricles^[4,5]. The second type comprises thin, elongated lipomas measuring less than 1cm in diameter, posteriorly situated curving around the splenium and are termed curvilinear. The curvilinear lipomas are generally associated with a normal or only mildly hypoplastic corpus callosum and otherwise have a low incidence of associated anomalies^[4,5]. The tubulo-nodular type is much more common.

In this article, I present a rare case of tubulonodular midline lipoma extending into both lateral ventricles to appear as a lipoma of choroid plexus associated with corpus callosum agenesis. In addition, I discuss the relevant pathogenesis which will possibly help to understand the morphological imaging behaviour of these malformations.

CASE REPORT

A 35-year-old man was referred to my hospital for investigation of epilepsy. Clinically the patient had emotional and psychological disturbance, otherwise no abnormality was seen on neurological examination. Radiological evaluation included magnetic resonance imaging (MRI) scanning. MRI images revealed lipoma occupied the area of the corpus callosum which was agenetic; continued through the choroidal fissure as bilaterally symmetrical lateral ventricular lipomas (Figures 1, 2). Pericallosal lipoma measured approximately

6.2 × 4.6 cm. Choroid plexus lipoma measured approximately 1.6 cm in maximum diameter. Bilateral symmetrical discrete areas of fatty tissue were noted in the cingulate sulcus. MR images also revealed agenesis of the corpus callosum and colpocephaly; the occipital, but not the frontal, horns of the lateral ventricles were dilated. The anterior cerebral artery and its branches were incorporated into the lipoma (Figure 3).

DISCUSSION

Intra-cranial lipomas are extremely rare, comprising 0.1 to 0.5 of all primary brain tumours. Pericallosal lipomas constitute 50 % of all intra-cranial lipomas and are associated with varying degrees of dysgenesis of corpus callosum^[6].

The pathogenesis of a pericallosal lipoma is considered to be the result of an abnormal resorption of the primitive meninges (meninx primitiva)^[7,8,9]. Usually, this resorption occurs between the eighth and the 10th week of development to create sub-arachnoid spaces. When the primitive meninges persists longer, instead of being resorbed, it differentiates into mature lipomatous tissue^[8,9]. Such lipoma may

develop in all the cerebral cisternae (hence, the lipomas of brain are mal-formations rather than a true neoplasm), but they are much more frequent in the area of the corpus callosum where it interferes with its normal growth between the 11th and 20th weeks^[8,9]. Therefore, anomalies of the development of the corpus callosum (complete or partial agenesis, hypoplasia) almost always coexist. The degree of anomaly seems to be in relation with the size and location of the lipoma. The normal sub-arachnoid nerves and vessels may also become traversed through these lipomas^[8,9].

As mal-development of primitive meninx occurs before the development of the inter-hemispheric commissural fissures, there is a frequent association of agenesis of corpus callosum with lipoma of CC^[8].

The development of lateral ventricular choroid plexus includes invagination of a portion of the inter-hemispheric cistern and tela choroidea of the ependymal roof plate through the choroidal fissure with infolding of the primitive meninx^[3,8,10]. The developing choroid plexus is attached to it and thus enters into the ventricle as well. In the absence of corpus callosum, pericallosal lipoma would use the same path to get into the lateral ventricles^[8]. This explains the association

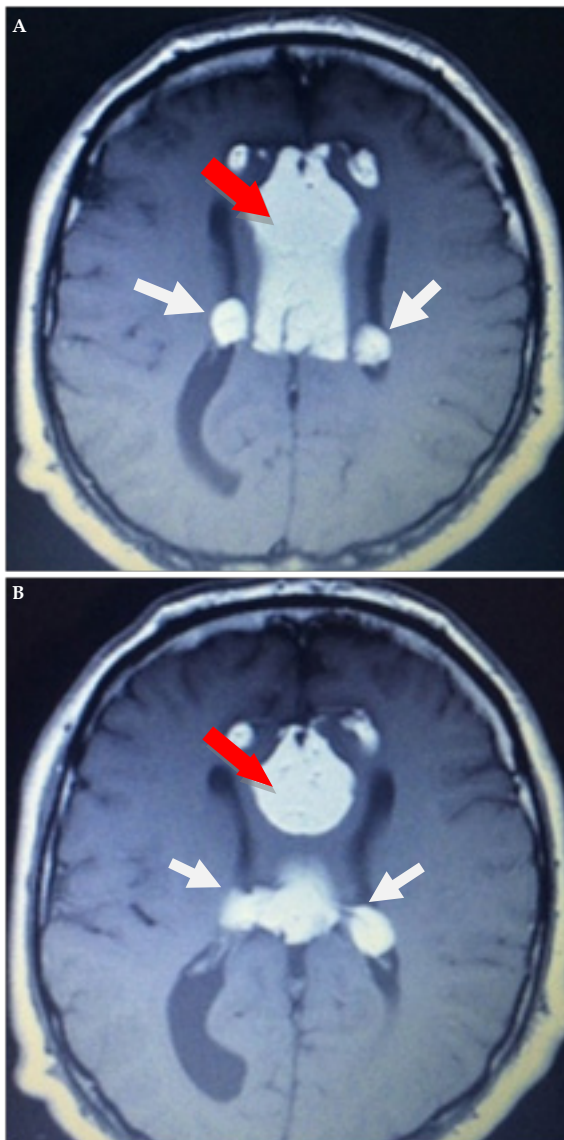


Figure 1 (A, B) axial T1 weighted MRI scans show pericallosal lipoma (red arrows) continuing as bilaterally symmetrical lateral ventricular lipomas of choroid plexus (white arrows).

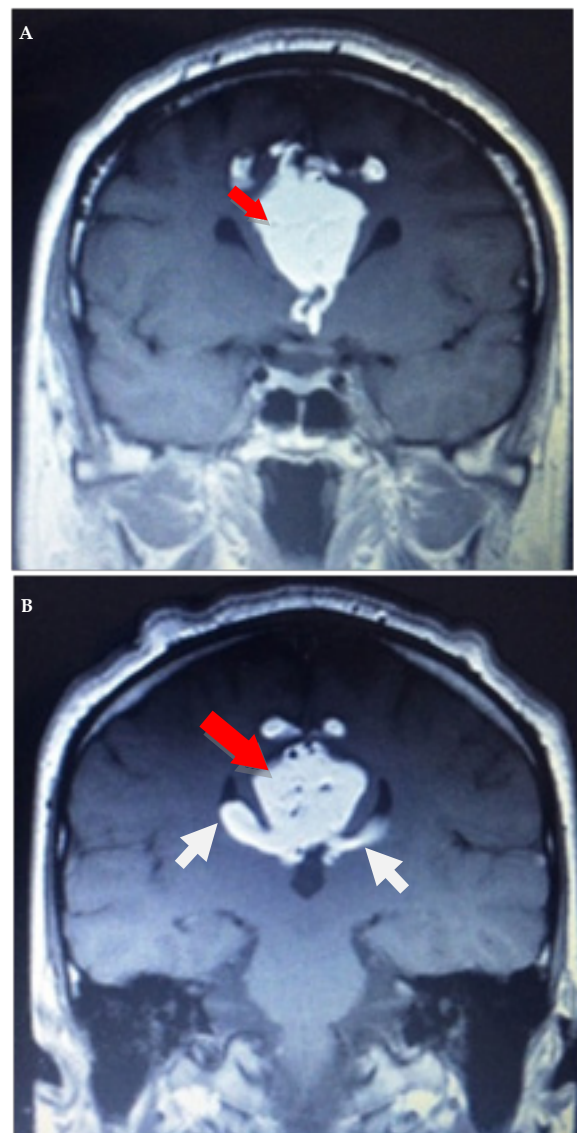


Figure 2 (A, B) Coronal T1 weighted MRI scans shows pericallosal lipoma with extension into both lateral ventricles through a narrow neck of choroid fissure to appear as a lipoma of choroid plexus.

of pericallosal lipoma with lipoma of choroid plexus in some cases. The incidence of this association been reported to be 20 to 50 %^[3,7].

In about 50 % of cases detection of pericallosal lipoma is

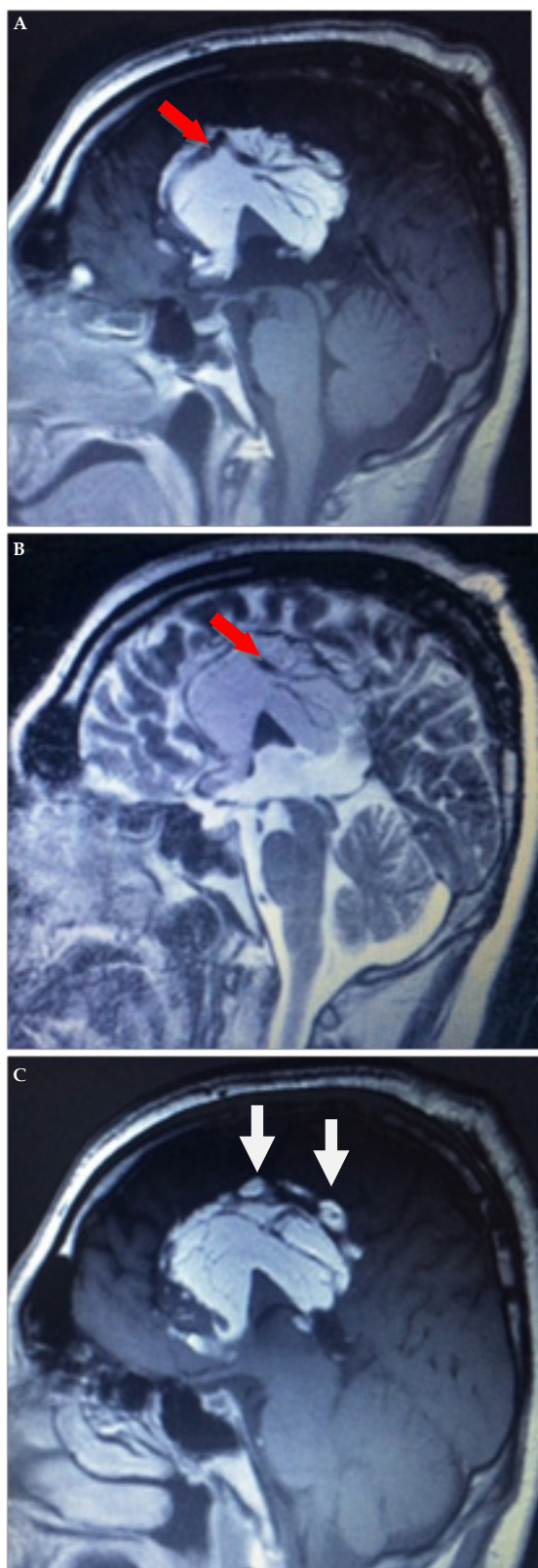


Figure 3 (A, C) Sagittal T1, (B) Sagittal T2 weighted MRI scans shows a high-signal-intensity (fatty) bulky mass in the midline of the anterior pericallosal area traversed by cerebral vessels (red arrows). Fat globules were scattered along the cingulate sulcus (white arrows), and agenesis of the corpus callosum was also evident.

incidental^[1,9]. The patients may present with seizures refractory to anticonvulsant treatment, headache, emotional lability, hemiparesis, mental retardation and psychological disorders. Our patient was investigated for epilepsy and he was apparently suffer from some emotional and psychological problems.

My patient had classical features of agenesis of corpus callosum: high riding third ventricle, separated and parallel lateral ventricles, colpocephaly and spoke wheel appearance of sulci over medial surface of cerebral hemispheres.

My patient had a large, tubulonodular midline lipoma having characteristic MRI signal intensities of fat with extension into both lateral ventricles through a narrow neck of choroid fissure to appear as a lipoma of choroid plexus. MRI as in my case was the modality of choice to easily appreciate this continuity because of the hyperintense nature of fat on T1WI in contrast to hypointense CSF, and also MRI was valuable for the characterization of the associated agenesis/dysgenesis of the corpus callosum. While in cases investigated by CT scan, the continuity is not easily depicted and the radiologist needs to change the viewing conditions and use wide windowing to appreciate slightly more hypodense fat from less hypodense CSF.

Because of their malformative origin, pericallosal lipoma usually do not become hyperplastic, whereas it might occur when the patient gains weight, as well as other sites of fat storage^[11].

No specific treatment for pericallosal lipoma is usually required^[1,9], although seizures if present need medical management and the prognosis depends on the presence of additional central nervous system abnormalities. Surgical therapy is usually not indicated because the risks of surgical intervention outweigh the potential benefits in most cases and may result in high morbidity and mortality rates, given the incorporated neurovasculature and strong adhesion to surrounding tissue^[12,13].

This case emphasize some concepts such as pericallosal lipoma associated with corpus callosum agenesis can extend into either or both lateral ventricles through choroid fissure to appear as lipoma of choroid plexus, and to appreciate the continuity or connection between the two lipomas, MR is superior to CT scan.

CONCLUSION

I report a rare case of a 35-year-old man with large tubulonodular pericallosal lipoma extending as lipoma of choroid plexus associated with agenesis of corpus callosum. To the best of my knowledge, this appearance has been described rarely in the published literature before. This rare exhibition of intracranial lipomas may advance new perception to anatomists or embryologists to verify or rebut different theories related to the origin of intracranial lipomas. Also, it is necessary for physicians and radiologists to know about this entity and its various atypical presentations to avoid diagnostic pitfalls and unnecessary investigations, which can have major consequences and complications for patients.

CONFLICT OF INTEREST

There are no conflicts of interest with regard to the present study.

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